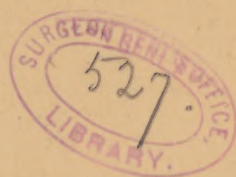


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A CASE OF PEMPHIGUS FOLIACEUS ENDING FATALLY  
WITHIN EIGHT MONTHS.

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THE history of the following case of pemphigus chronicus vulgaris, turning into pemphigus foliaceus and ending fatally within about eight months, has been taken from my notes, principally at the suggestion of my friend, Dr. J. W. Gleitsmann, Professor of Laryngology in the New York Polyclinic, on account of its bearing on the incidental affections of the oral cavity and the larynx. Dr. Gleitsmann having abandoned the idea of writing a paper on the subject, the case, I believe, is of interest enough in itself to justify its publication, since only very few cases of pemphigus foliaceus have been recorded in this country, those of Dr. Sherwell, of Brooklyn, and of Dr. Graham, of Toronto, Canada, mentioned by Duhring, being the only ones I could find after a careful search of the *Index Medicus*. Besides, interest in the case will be somewhat increased by the fact that recently the pathology of pemphigus was brought into greater prominence by the discussion of the subject before the late meeting at Vienna of the German Congress for Internal Medicine, in connection with a report by Mosler, of Greifswald, in which the participation of the mucous membranes formed an important part. The present case has been briefly mentioned by Dr. L. D. Bulkley in a paper published recently.<sup>1</sup>

Mr. F. T., cigar manufacturer; a native of Germany; forty-nine years of age, with a good family record; married, and father of six healthy children. Was first seen by me at my office on May 7, 1887. A man of strong physique, he had generally enjoyed good health; he at one time was a member of the police force of this city; he never had syphilis; in 1876 he had pneumonia, and, somewhat later, trouble with the left tonsil, which he subsequently had removed. In December, 1886, he contracted a severe cold at the inauguration of the Montefiore Home, and on another occasion soon afterward had his feet thoroughly chilled. Within a short time he began to suffer with bronchitis. In January, 1887, he noticed a blister on the left breast, and soon after blisters in the mouth, particularly on the gums, which caused a great deal of annoy-

<sup>1</sup> New York Med. Journal, 1889, i., No. 15: "On the Value of Frequently Repeated Doses of Arsenic in Treatment of Bullous Diseases of the Skin, etc."





ance, especially during eating and otherwise. He consulted several specialists for mouth and throat diseases, without relief. It appears that the lesions in the oral cavity were considered as specific, and were treated accordingly. I saw the patient first in consultation with his family physician, Dr. J. Scheider, and with Dr. Gleitsmann, because the latter had some doubt as to the nature of the lesions of the skin as well as of the mouth.

On inspection of the oral cavity the gums, from the edge about half-way to their base, appeared devoid of epithelium, leaving a dark-red, nearly cherry-colored surface, with intervening yellow spots here and there. The mucous membrane of the cheeks, of the uvula, and of the soft palate showed the same loss of the epithelial cover, forming irregular and variously outlined dark-red patches, partly covered with yellowish membranes resembling mild diphtheritic exudations. Between the upper and lower maxillæ this condition was particularly severe, shallow fissures of a deep-red color appearing on the generally yellowish surface. On the upper surface of the tongue, as well as on the lower one, numerous dark-red raw spots were likewise noticed.

On the scalp, which, particularly in the front parts, was but thinly covered with dark-brown hair, a number of bullæ of different sizes, more or less tensely distended with a clear, watery fluid, were observed; the largest one, about the size of a fifty-cent piece and rather flat, was located just within the hair, several smaller ones over the vertex, and some on the forehead. Beside these blebs a number of pale reddish spots could be noticed on the face and scalp, some of them quite smooth, some covered with thin scales resembling gold-beater's skin. On the right aspect of the neck a number of similar bullæ were found, one behind the left ear covered with a blackish crust owing to the application of nitrate of silver, and further down on the same side a larger cluster of smaller bullæ or vesicles containing a perfectly clear fluid.

Quite a number of similar bullæ and scaly or smooth spots were scattered over nearly the entire front and back of the thorax without forming distinct groups, while on the abdomen only a few blisters could be observed, besides numerous reddish spots, which, according to the patient's observation, mark the first stage in the development of blisters. The bullæ vary in size from a hemp-seed to a small filbert; the more recent ones are all hemispherical and tensely distended, while some of the older ones are flaccid and overlap their bases like a bag. The contents of the blebs were in almost all cases perfectly clear, only in the dependent portions of some of the older ones a somewhat opaque or even pus-colored fluid could be observed. Close to the frenulum glandis penis two small excoriated depressions of a remarkably dark-red color, resembling that of raw smoked beef, were found; on no other part of the body, particularly not on the extremities, could blisters or scars or other pathological changes be detected; the lymphatic ganglia were nowhere enlarged or otherwise affected.

On the strength of these symptoms, in connection with the condition of the mouth, and with the history of their development, the diagnosis of pemphigus chronicus vulgaris was made. The patient was ordered to take arsenic and iron (Fowler's solution with the tinctura ferri pomati, 1 part to 12; 20 to 30 drops three times a day); locally, baths and a dusting powder composed of bismuth, zinc, and starch were given. Considering the comparatively small extent of the disease after a dura-

tion of over three months, it could be regarded as of a mild type. Up to May 11th very few new blebs had made their appearance, except in both axillæ, where clusters of smaller bullæ were visible, some containing a transparent fluid of a light-yellow color; others were dried up into a yellowish crust. The maculæ on the abdomen had entirely disappeared.

*May 17th.* No new blebs were found; almost all the former ones had dried up, leaving brownish spots, some of them scaly. Even the axillary eruption showed great improvement, but the excoriations on the penis had not undergone any change. The dose of the medicine had been increased to 40 drops without any trouble, except slight costiveness.

*25th.* While only a few new bullæ have broken out on the body, numerous clusters of smaller ones have developed between the toes and on the soles of both feet, while the large one on the scalp, as well as another quite conspicuous one over the right clavicle, have entirely dried up. The nail of the right thumb shows a brownish color, suggesting the formation of a blister beneath that had rapidly dried up again. The patient's general health is fair; he has retained the weight of one hundred and seventy-two pounds to which he had previously fallen.

*June 1st.* A few new bullæ on the right arm and the thorax have appeared, but quite numerous ones beneath and between the toes which are very annoying on account of itching and of the offensive smell of the easily decomposing contents. After removing the cuticula as far as possible with a pair of curved scissors, the parts were cleansed with a solution of boric acid, dusted with bismuth, and covered with borated cotton.

In this way the disease continued, taking a still comparatively mild course, up to June 20th; although constantly new blisters formed, some of them in groups, still they had the tendency of drying up rapidly and to leave the skin but little impaired, except as to its color, which was changed into a reddish-brown. From that date on, however, it assumed a different character. Not only did the isolated bullæ all over the trunk become more numerous, but also, first on the right forearm and very soon afterward on the left one, more flaccid bullæ appeared in great numbers; they extended within a few days from the shoulders to the wrists and to the backs of the hands, and soon began to run together, detaching the epidermis over a considerable space in the centre and leaving a denuded, bright-red, shining or glazed surface, as in eczema rubrum, or that beneath the blisters of a burn of the second degree. Although in the central portions of such patches new epidermis was formed, it never became solid and firm again, but remained brittle, divided by superficial fissures, or slightly scaling. In the peripheral portions, particularly around the wrists, the bullæ lost their sharply defined form and the epidermis was raised in a continuous outline, clearly exhibiting the character of pemphigus foliaceus.

After June 20th the patient was unable to dress, and had to stay at home. Arsenic, the dose of which had heretofore been gradually increased, was now omitted, owing to loss of appetite and dyspeptic symptoms, and 5 grains of the muriate of quinine were given every three hours. Local baths were applied to the arms; but on the 22d the eruption of bullæ became so general all over the trunk that the patient was ordered to stay in the bath-tub as continually as possible at a temperature of 80° to 85°. This he did from that date with little



interruption up to August 1st, generally repairing into the bath-tub at 5 or 6 o'clock A.M., and remaining there as late as 9 P.M., taking all his meals while there. The warm weather prevailing throughout July, 1887, favored the use of a bath as continuously as circumstances would permit. During the night lotions or powders were applied, and the compound salicylated soap plaster which I have devised<sup>1</sup> was used on the larger raw patches with good effect.

A slight remission followed this first stormy eruption, lasting, however, but a few days. The bullæ now numbered by hundreds, and new ones were constantly added, while the old ones rapidly dried up and even the denuded surfaces showed considerable improvement. Except a feeling of weakness the general condition of the patient was fair, appetite good, bowels regular; sleep at night was not very quiet, but the patient had a number of naps during the day. The urine at this time and through the entire course of the disease never showed any irregularities; it was voided, without any trouble, in normal quantities, was generally of a light-yellow color, and never showed the presence of albumin or sugar.

A new feature manifested itself after June 27th. Heretofore all the new blebs appeared on skin of normal condition (except on the arms, where it was considerably swollen and red during the intense eruption), and had caused but slight burning or itching preceding the eruption. On the above date it was noticed that some of the bullæ on the trunk rose on top of a wheal-like, reddish elevation, accompanied by considerable itching. On the next day numerous slightly elevated patches of a dusky-red color were observed on the lower part of the trunk, extending nearly symmetrically to the thighs, varying in size from a silver half-dollar to a dollar. Over these patches numerous small blisters with perfectly clear contents could be observed, principally studding the borders of the patches, most of them yet half-hidden in the skin, causing intense itching before and after eruption. This condition answered very well to the description of pemphigus pruriginosus given by Kaposi.

While the general condition of the patient remained satisfactory, on June 30th it was noticed that for the first time bullæ appeared on parts previously unaffected; on the back and abdomen several blisters attained the size of a walnut, raised more than half an inch above the surface of the skin. On July 1st the patient was seen in consultation by Dr. George H. Fox, who proposed mainly a change of diet to a more vegetable and less stimulating one.

It is no longer necessary to follow so closely the course of the disease. During the first part of the month of July there followed a general improvement. Still, new blisters were forming constantly and on some localities large areas denuded of epidermis could always be found, while in certain portions, particularly around the joints—above all, about the wrists—the foliaceous character never entirely disappeared. The patches, resembling urticaria, gradually flattened down and coalesced, forming extensive areas of a dark, bluish-red color, which, at first beginning only around the borders, soon changed into a brighter, almost scarlet, tint, and presented its former deep hue only after the patient had assumed an upright position for some time. The groups of smaller vesicles, often half-hidden in the skin, with their intense itching,

<sup>1</sup> New York Med. Journal, Sept. 17, 1887.

continued to appear beside the isolated larger bullæ arising from the uninfiltreated skin. After July 6th the quinine was omitted and 10 grains of salicylate of soda were given every three hours. This drug seemed to have a beneficial effect on the itching and on the formation of the smaller blisters, but the general condition was not much altered; therefore, on the 17th Fowler's solution was again resorted to, increasing from 2 to 5 drops three times a day.

About July 19th an extensive eruption over the buttocks obliged the patient to relinquish the bath-tub for several days, as the pressure on these parts became unbearable; but he soon returned to the bath, as apparently he found more comfort in the water than in the bed. About the same time the affection of the mouth and lips, which so far had continued in a moderate degree, began to cause intense suffering, and obliged the patient to avoid solid food almost entirely. Dr. D. L. Bulkley, who saw the patient for the first time on July 26th, recommended a more effective administration of arsenic, which now was tried in the form of the solution of sodium arsenatis, 2 to 10 drops every two hours.

A short period of improvement set in again about August 1, although the formation of new blisters did not cease entirely, and several portions remained at all times severely affected; in general the tendency to form a new epidermis decidedly prevailed over that of destruction. The affection of the mucous membrane of the mouth and its adnexa, however, remained exacerbated, and considerably annoyed the patient by the discharge of a copious serous secretion, which originated either in the pharynx or the œsophagus or the trachea or the larger bronchi.

Heretofore the temperature had always remained normal and the pulse seldom exceeded 72. On the evening of August 5th Dr. Scheider, who continuously attended Mr. T., found the temperature at 101°, pulse 120, with frequent respiration, and the patient in a state of great weakness. The next day at noon, with a temperature of 102°, pulse 112, respiration 30, he was very low and weak, continuously expectorating large quantities of a sero-purulent fluid from the throat. The symptoms suggested congestion of the lungs or beginning pneumonia, of which, however, no physical signs could be detected by careful examination. Under the administration of valerian with the carbonate of ammonia and stimulants, the temperature remained between 101° and 100.5°, the pulse about 96, the respiration somewhat more frequent than normally for the next three days. After August 10th the temperature again became normal, the pulse less frequent, the secretion gradually less, and the general condition of the patient improved considerably, leaving him, however, a great deal weaker physically and psychically much more depressed. He repaired to the bath-tub only occasionally and for short periods on account of increasing weakness, although he longed for the relief afforded by the bath.

Throughout the remainder of August the disease continued its course with little change; the temperature showed occasional elevations to 100.8° and 101.4°; the pulse retained a frequency of 90 beats and over. While the patient was able to take sufficient quantities of nourishment, as the mouth and throat were in much better condition again, and the secretion therefrom had almost entirely ceased, the bowels exhibited a tendency to looseness, three to five passages a day being the rule. The local process on the skin showed occasional remissions, but short and slight. From the time the bath was given up the scaling became much



more conspicuous, and on some parts considerable accumulation of the brittle, grayish or yellowish scales took place, often hiding from view the real condition of the denuded corium underneath and exhibiting more distinctly that condition to which the name of pemphigus foliaceus owes its origin. At the same time a peculiar smell, mentioned by several authors, became noticeable, to gradually give way to the more decided offensive odor of decomposing epidermis. Altogether the patient presented a most pitiable sight: new blisters were constantly appearing all over the body, repeated eruptions taking place on the same localities, and hardly a square inch of normal skin could be found. Now the inguinal, now the popliteal region, now penis and scrotum, now neck and back, were the parts principally affected, generally showing the corium exposed over a large area, with bleeding and oozing fissures wherever the skin is exposed to flexion and extension; even the face became affected extensively, particularly the chin, lips, and eyelids, which at different times were entirely devoid of epidermis or covered with black crusts. The conjunctiva of the lids and of the bulbs blistered in their turn, and sores remained on the swollen mucous surface for some time. Around the heels the blistering process became extremely painful, owing in part to the thickness of the epidermis, which could not be detached so easily, and with its sharp, hard edges cut deep, extremely sensitive fissures. To relieve the local lesions, lotions, powders, ointments, plasters, and cotton padding were employed wherever it was found possible to do so, but scarcely had the last spot been attended to when the first ones again required attention, so that there was not a moment of real rest or comfort. Arsenic was continued as regularly as possible, together with opiates to check the diarrhoea.

About the 1st of September the mouth, throat, and larynx again became worse, while externally several places began to dry up. Owing to my absence from New York, I did not see the patient again after that date. From personal communications from the attending physician, Dr. Jul. Scheider, and from notes taken by the nurse, I shall briefly report the final course of the disease: The rise of temperature was more constant and more pronounced, reaching  $102.4^{\circ}$  on the 2d and again on the 5th of September, after a slight remission; it attained  $103^{\circ}$  on the 6th, to fall again to  $102.6^{\circ}$  on the following days. The pulse became much more frequent: after rising to 108 on September 4th, it increased to 128 on the 5th, 130 on the 6th, 136 on the 7th, and continued about 120 to the end. Diarrhoea became much more profuse from day to day, six passages occurring on the 1st of September, eight on the 2d, nine on the 3d, ten on the 4th and 5th, twelve on the 6th, twenty-one on the 7th, 8th, and 9th. The patient gradually lost strength, without ever losing consciousness, and died on the morning of September 10th. During the last few days, with the rapid increase of diarrhoea, no more new blisters appeared.

There is a certain temptation to range this case among the cases of dermatitis herpetiformis on account of the intercurrent appearance of the clusters of small, very itchy vesicles on hyperæmic patches, which, indeed, were more or less herpetiform, but otherwise the disease did not exhibit the characteristic features of dermatitis her-



petiformis. In the later stages the disease had much in common with impetigo herpetiformis, the contents of the bullæ being purulent from the start, in many instances; but for a long period there existed only bullæ with clear, serous contents and no pustules at all, thus distinctly differing from impetigo. Unfortunately, no autopsy could be obtained, which certainly would have elucidated one point in which I was particularly interested—the origin of the diarrhœa. I have little doubt that it was due to the extension of the pemphigoid process (of forming bullæ and destroying the epithelial cover) to the intestinal tract. There is certainly some probability of such an occurrence in view of the participation of the mucous membranes of the oral cavity, the larynx, and the œsophagus; and this supposition finds a valuable support in the observation in Dr. Graham's case, where the report on the autopsy says:<sup>1</sup> "Mucous membrane along the lesser curvature of the stomach was congested, and about twenty spots were discovered which nearly resembled some of the patches of eruption on the skin. The spots varied in size from that of a pea to a ten-cent piece, and on close inspection were found to consist of superficial ulcerations, some extending partly and some altogether through the mucous membrane. The mucous surface of the large intestine was very much congested and thickened in some places. Death seems to have been hastened by the eruption, so to speak, on the mucous surface of the stomach."

Concerning the effects of treatment, I must candidly confess that I do not believe the course of the disease was at all influenced by the same. There were, indeed, several periods of improvement, but I consider them rather as spontaneous fluctuations—exacerbations and remissions being more or less the rule in all cases of chronic pemphigus. I therefore cannot agree with Dr. Bulkley as to the effectiveness of arsenic, certainly not in the case of Mr. T., although I am well aware that in a number of cases it has undoubtedly been given with great benefit and is considered almost the only remedy worthy of a trial. It was given early and later on certainly in abundant doses, but, as the final result proved, without real effect. I cannot suppress a suspicion that arsenic had something to do with the profuse passages, although I cannot bring forth any positive evidence in favor of this opinion. Generally, arsenic shows its best results in skin diseases accompanied by dryness and scaling, rather where an excessive formation of the corneal layer of the epidermis takes place. Why it should have the same effect in a disease where the formation of epidermis is defective, rather, and a constant loss of the same takes place, as in pemphigus, is not easily understood. Although we have lately heard less of the advantages of the internal

<sup>1</sup> Canadian Journal of the Medical Sciences, 1879, p. 172 et seq.

use of ichthyol as proclaimed by Unna, I should feel inclined to try it whenever I may meet with a similar case again.

The external applications were made with the purpose rather of relieving the suffering of the patient than of effecting a cure. There is no doubt in my mind that the use of the protracted bath gave the greatest relief, and that it ought to be resorted to wherever practicable.

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